


● **DIRECTIONS FOR ADMINISTRATION** Powder may be mixed with drinks or taken as a paste.

● **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug. Forms available from special-order manufacturers include: tablet, oral solution, powder

Oral solution

▶ Citrulline (Non-proprietary)

L-citrulline 100 mg per 1 ml Stimol oral solution 10ml sachets sugar-free | 36 sachet [PoM] 

Powder

▶ Citrulline (Non-proprietary)

L-citrulline 1 mg per 1 mg L-Citrulline powder | 100 gram £249.99

BENZOATES

Sodium benzoate

10-Mar-2020

● INDICATIONS AND DOSE

Acute hyperammonaemia due to urea cycle disorder (specialist use only)

▶ BY INTRAVENOUS INFUSION

▶ Neonate: Initially 250 mg/kg, to be administered over 90 minutes, followed by 10 mg/kg/hour, adjusted according to response.

▶ Child: Initially 250 mg/kg, to be administered over 90 minutes, followed by 10 mg/kg/hour, adjusted according to response

Maintenance treatment of hyperammonaemia due to urea cycle disorders (specialist use only) | Non-ketotic hyperglycaemia (specialist use only)

▶ BY MOUTH

▶ Neonate: Up to 250 mg/kg daily in 3–4 divided doses, dose to be taken with feeds.

▶ Child: Up to 250 mg/kg daily in 3–4 divided doses, dose to be taken with feeds or meals; maximum 12 g per day

● **UNLICENSED USE** Not licensed for use in children.

● **CAUTIONS** Conditions involving sodium retention with oedema (preparations contain significant amounts of sodium) · congestive heart failure (preparations contain significant amounts of sodium) · neonates (risk of kernicterus and increased side-effects)

● **SIDE-EFFECTS** Appetite decreased · coma · irritability · lethargy · nausea · vomiting

SIDE-EFFECTS, FURTHER INFORMATION Gastro-intestinal side-effects may be reduced by giving smaller doses more frequently.

● **PREGNANCY** No information available.

● **BREAST FEEDING** No information available.

● **RENAL IMPAIRMENT** Caution in renal insufficiency—preparations contain significant amounts of sodium.

● DIRECTIONS FOR ADMINISTRATION

▶ With oral use For administration *by mouth*, oral solution or powder may be administered in fruit drinks; less soluble in acidic drinks.

▶ With intravenous use For *intravenous infusion*, dilute to a max. concentration of 50 mg/mL with Glucose 10%.

● PATIENT AND CARER ADVICE

Medicines for Children leaflet: Sodium benzoate for urea cycle disorders www.medicinesforchildren.org.uk/sodium-benzoate-urea-cycle-disorders

● **MEDICINAL FORMS** Forms available from special-order manufacturers include: tablet, capsule, oral solution, solution for infusion

DRUGS FOR METABOLIC DISORDERS > ACETIC ACIDS

Sodium dichloroacetate

● INDICATIONS AND DOSE

Pyruvate dehydrogenase defects

▶ BY MOUTH

▶ Neonate: Initially 12.5 mg/kg 4 times a day, adjusted according to response, increased if necessary up to 200 mg/kg daily.

▶ Child: Initially 12.5 mg/kg 4 times a day, adjusted according to response, increased if necessary up to 200 mg/kg daily

● **SIDE-EFFECTS** Metabolic acidosis · oxalate metabolism abnormal · polyneuropathy (long term use)

● **PREGNANCY** No information available.

● **BREAST FEEDING** No information available.

● **MEDICINAL FORMS** No licensed medicines listed.

DRUGS FOR METABOLIC DISORDERS > AMMONIA LOWERING DRUGS

Glycerol phenylbutyrate

13-Sep-2018

● **DRUG ACTION** Glycerol phenylbutyrate is a nitrogen-binding agent that provides an alternative vehicle for waste nitrogen excretion.

● INDICATIONS AND DOSE

Urea cycle disorders (specialist use only)

▶ BY MOUTH, OR BY GASTROSTOMY TUBE, OR BY NASOGASTRIC TUBE

▶ Neonate (body surface area up to 1.3 m²): Initially 9.4 g/m² daily in divided doses, usual maintenance 5.3–12.4 g/m² daily in divided doses, each dose should be rounded up to the nearest 0.1 mL and given with each feed. For dose adjustments based on individual requirements—consult product literature.

▶ Child 1–23 months (body surface area up to 1.3 m²): Initially 9.4 g/m² daily in divided doses, usual maintenance 5.3–12.4 g/m² daily in divided doses, each dose should be rounded up to the nearest 0.1 mL and given with each meal or feed. For dose adjustments based on individual requirements—consult product literature

▶ Child 2–17 years (body surface area up to 1.3 m²): Initially 9.4 g/m² daily in divided doses, usual maintenance 5.3–12.4 g/m² daily in divided doses, each dose should be rounded up to the nearest 0.5 mL and given with each meal. For dose adjustments based on individual requirements—consult product literature

▶ Child 2–17 years (body surface area 1.3 m² and above): Initially 8 g/m² daily in divided doses, usual maintenance 5.3–12.4 g/m² daily in divided doses, each dose should be rounded up to the nearest 0.5 mL and given with each meal. For dose adjustments based on individual requirements—consult product literature

DOSE EQUIVALENCE AND CONVERSION

▶ 1 mL of liquid contains 1.1 g of glycerol phenylbutyrate.

▶ For patients switching from sodium phenylbutyrate or sodium benzoate—consult product literature.